Wilms Tumor With Metastasis to the Vagina: A Case Report

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A 12-year-old female presented with abdominal pain, night sweats, weight loss, constipation, dysmenorrhea, menorrhagia, and vaginal discharge. Examination revealed a palpable flank mass and a large tumor adherent to the anterior vaginal wall. Computed tomography scan demonstrated a 23 cm mass in the left kidney, a separate 10.8 cm pelvic mass, and metastatic disease. Biopsies were consistent with Wilms tumor. Neoadjuvant chemotherapy and a left radical nephrectomy were performed for her stage IV disease as the kidney was amiable to complete resection. The patient received radiation and resumed chemotherapy. She was doing well with improved symptoms at follow-up.

CASE REPORT

A 12-year-old female presented to the emergency department with worsening abdominal pain, drenching night sweats, a 10 lb weight loss over the past month, constipation, urinary urgency and incontinence, weak stream, dysmenorrhea, menorrhagia, and vaginal discharge. She was recently treated for bacterial vaginosis. The patient was otherwise healthy without medical or surgical history, significant family or social history, and was not taking medications or supplements. Her physical examination found her afebrile with stable vital signs, a palpable and firm left flank mass, and a large protruding tumor adherent to the anterior vaginal wall. The patient had normal external female genitalia and no palpable adenopathy. Laboratory evaluation demonstrated white blood count of 14.7, Hgb of 8.0, hematocrit of 26.1, platelets of 563, Cr of 0.74, albumin of 3.1, and electrolytes that were within normal range, as were her serum human chorionic gonadotropin and alfa fetoprotein. Urinalysis showed red and white blood cells, and urine culture had no growth. An abdominal ultrasound revealed a complex heterogeneous mass with hyperechoic elements measuring 25 cm in the left retroperitoneum and distinct solid homogeneous pelvic mass hypoechoic cystic components. Computed tomography scan of the chest, abdomen, and pelvic with contrast demonstrated a cystic components. Computed tomography scan demonstrated a 23 cm mass in the left kidney, a separate 10.8 cm pelvic mass, and metastatic disease. Biopsies were consistent with Wilms tumor. Neoadjuvant chemotherapy and a left radical nephrectomy were performed for her stage IV disease as the kidney was amiable to complete resection. The patient received radiation and resumed chemotherapy. She was doing well with improved symptoms at follow-up.

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Postoperatively, the patient underwent a stem cell harvest and received whole body irradiation to the chest, abdomen, inguinal region, and pelvis of 2100 cGy in 14 fractions. She resumed chemotherapy with the addition of cyclophosphamide and etoposide per regimen M as she had improved but persistent disease in the liver and lung. The vaginal mass had decreased to 4.5 cm. The patient was doing well with improved symptoms at 4 months postoperatively, tolerating chemotherapy, voiding and stooling well, and without pelvic symptoms. Therefore, decision was made to continue observation of the vaginal lesion and hold off on debulking surgery for now.

DISCUSSION
Wilms tumor is classically a malignant tumor of the kidneys composed of metanephric blastema, stromal, and epithelial
elements seen in children. To our knowledge, this is the first report of a vaginal mass presenting as metastatic Wilms tumor. When abnormal vaginal findings have been reported in the literature with Wilms tumor, they are in relation to a congenital anomaly (septation, duplication, benign tumors, etc) and are usually associated with mutations in the WT-1 gene or Denys-Drash syndrome.27

Isolated Wilms tumor of the female reproductive tract has been reported in rare case reports. A 13-year-old girl who presented with vaginal spotting had a vaginal Wilms tumor composed of triphasic tissue with primitive cartilage and skeletal muscle with squamous and columnar epithelium. After surgical excision and chemotherapy, the patient was reported to be disease-free for over 7 years.8 Wilms tumor was reported to arise from the cervix of a 12-year-old girl presenting with vaginal discharge. Biopsy of a bulky mass confirmed Wilms tumor with favorable histology; chemotherapy with vincristine, doxorubicin, cyclophosphamide, carboplatin, and etoposide led to significant reduction of the mass facilitating transection at the stalk and cold-knife conization of the cervix yielding her in complete remission at 12 months.9 Another 13-year-old girl with isolated cervical Wilms presented with a polypoid mass attached to the endocervix by a stalk. The biopsy showed blastemal, epithelial, and stromal elements. Although the patient was not given organ-sparing therapy, she is reported to be disease-free for over 9 years after hysterectomy.10

Wilms tumor of the uterus was first reported in 1974 in a 14-year-old girl admitted with abdominal cramping and vaginal bleeding since menarche 1 year prior. Physical examination showed a large tumor involving the vagina and pouch of Douglas. Laboratory examination was normal aside from moderate anemia. The patient was treated with hysterectomy with culdotomy followed by adjuvant chemotherapy, and remained disease-free after 5 years of follow-up. Histologic examination of the tumor showed embryologic tubules, glomeruli, and blastema within a myxomatous stroma.11 Wilms tumor has also been seen in the uterus of patients as old as 44 years, such as one presenting with a bleeding polypoid cervical mass, and after hysterectomy followed by chemotherapy was disease-free at 1 year.12 Uterine Wilms has been noted in 3 other patients, 2 adolescents as polypoid masses protruding through the cervix, and a 22-year-old presenting with menometrorrhagia. Triphasic differentiation with mesenchymal cells showed rhabdomyoblastic and epithelial differentiation, and most of the tumors was composed of blastemal cells.13

CONCLUSION

Extrarenal Wilms is extremely rare but appears to clinically behave similarly to renal Wilms. This is the first report of a vaginal mass presenting as a metastasis of renal Wilms tumor. The clinical course and prognosis of Wilms metastatic to the vagina have yet to be determined.

References